esophageal diameter and the small intestinal clearance (SIC) (EULAR 2011). We also suggested that anti-centromere antibodies (ACA) are an important factor of esophageal dilatation in SSc patients (EULAR 2010). However, changes of small intestinal involvement during long-term follow-up have not yet been defined.

Objectives: The aim of this study was to evaluate a correlation between the small intestinal involvement and clinical features in SSc patients during ten years follow-up.

Methods: Fifty-five patients with a definite diagnosis of SSc [52 female and 3male with mean age of 59.4 years (range 29–77 years)] were included in the study. Thirteen (23.6%) patients were classified as diffuse SSc and 42 (76.4%) as limited SSc. The SIC grade was classified according to barium meal reach at 30 min after intake; grade 1 (>2/3 of the whole small intestine), grade 2 (1/3~2/3), grade 3 (<1/3), grade 4 (the duodenum). The SIC change was classified as follows; grade 1 and 2 or grade non-increase during follow-up was classified as "non-progressive", grade 3 and 4 or grade increase as "progressive". All SSc patients were evaluated the items as used for 2013 ACR/EULAR criteria.

Results: The mean durations of follow-up period were 9.5±0.6 years. The number of SSc patients in each SIC grade at the initial evaluation and the end of follow-up were as follows; grade 1: initial vs end; 23 vs 24, grade 2: 20 vs 9, grade 3: 10 vs 16, grade 4: 2 vs 6. The SIC change "progressive" was 40.0% of SSc patients. Positive correlation between the esophageal diameters and the SIC grade was observed in SSc patients at the initial evaluation ($r=0.61$ $p<0.01$) and the end of follow-up ($r=0.71$ $p<0.01$). The esophageal diameters at the initial evaluation were significantly wider in SIC "progressive" group than in "non-progressive" group (non-progressive vs progressive; 21.8±6.5 vs 30.9±8.6 mm, $p<0.0001$). The frequencies of SSc patients with ACA-positive and sclerodactyly were higher but with puffy fingers were lower in SIC "progressive" group than in "non-progressive" group (non-progressive vs progressive; 24.2 vs 59.1%, p=0.009; 33.3 vs 63.6%, p=0.03; 72.7 vs 45.5%, p=0.03). The prevalence of pitting scar and Interstitial lung disease were tended to be higher in SIC "progressive" compared to SIC "non-progressive" group (non-progressive vs progressive; 15.6 vs 36.4%, p=0.07; 21.1 vs 45.5%, p=0.06).

Conclusions: The present study demonstrated that the progression of small intestinal involvement was associated with esophageal dilatation at the initial evaluation during long-term follow-up of SSc patients. Our findings also suggested that ACA and skin thickening of the fingers were important factors of small intestinal involvement in SSc patients. The SIC may be a useful tool for the assessment of GI tract involvement in SSc patients during long-term follow-up.

Disclosure of Interest: None declared

DOI: 10.1136/annrheumdis-2017-eular.2353

OP0128 DETECTION OF SUB-CLINICAL DIFFUSE MYOCARDIAL FIBROSIS BY NATIVE T1 MAPPING MAGNETIC RESONANCE IMAGING IN A PROSPECTIVE SYSTEMIC SCLEROSIS COHORT

V. Poindron, E. Chatelus, M. Canuet, P. Germain, S. El Ghannudi, T. Martin.
National Referral Center for Systemic Autoimmune Diseases, Hôpitaux Universitaires de Strasbourg, Strasbourg, France

Background: Cardiac involvement in systemic sclerosis (SSc) is the second most frequent SSc-related cause of death. It remains mostly asymptomatic in early stages and is underdiagnosed with routine non-invasive screening. Cardiac magnetic resonance imaging (CMR) is becoming a key actor as it has a better sensitivity than echocardiography (echo). CMR can detect diffuse myocardial fibrosis (DMF) by native T1 mapping, myocardial edema (ME) by T2 mapping and focal fibrosis by late gadolinium enhancement (LGE). Ntusi *et al.* reported an increase of 5% of T1 value suggestive of DMF in 10 of 19 SSc patients[1].

Objectives: To determine the prevalence of cardiac involvement by CMR native T1 and T2 mapping and its correlation with echo data and non-cardiac manifestations in SSc patients.

Methods: 72 patients fulfilling ACR/EULAR classification criteria were prospectively included between 2013–2016. They underwent CMR at 1.5T, including native T1 and T2 mapping, and LGE. Normal T1 value determined in our center was 1032±39 msec. In the present study an elevated native T1>1082 msec was likely representing ventricle (especially left) DMF and T2>55 msec representing ME [2].

Results: Patients characteristics: mean age: 56±14.8; diffuse disease: 38 (52.8%); anti-Scl70 positivity: 29 (40.3%); anti-RNApolIII positivity: 6 (8.3%); 21 (29.2%) patients had early disease (<2 years from first non-Raynaud symptom). The mean T1 was 1064±41.6 msec and T2 was 51.8±2.9 msec. 36 patients (50%) had DMF but only 6 (8.3%) had ME. The mean T1 in DMF cases was 1097±14, and the T2 in ME cases was 58.2±1.6 msec. LGE was reported in 25.7% of patients. Although LGE was more frequent in patients with DMF than in those without DMF (13 vs 5, p=0.024), only 13 (36.1%) DMF patients had LGE. Left ventricular ejection fraction (L-VEF), left ventricular telediastolic volume (L-VTDV), Right-VEF and Right-VTDV were similar in DMF and non-DMF (N-DMF) groups. Echo was normal in 18 (50%) patients with DMF and in 25 (69.4%) of N-DMF group (p=0.09). DMF and N-DMF groups were similar for sex ratio, age, cardiovascular risk factors and ischemic heart disease. DMF was more frequent in patients with late disease (27 vs 9, p=0.05). T1 value was positively correlated to pulmonary arterial hypertension and digital ulcerations together ($r=0.31$, p=0.008) but not with Rodnan skin score. Six patients (8.3%) died during the inclusion period: 5 were in DMF group (p=0.09). The alterations of L-VEF and R-VEF were correlated ($r=0.45$, p=0.009). DMF was not associated with skin subsets, interstitial lung disease, auto-antibody profile, all echo parameters, CRP and BNP.

Conclusions: Native T1 mapping detects left ventricular DMF in 50% of patients with SSc including 43% (9/21) of the patients with early disease. Among them, 36% had normal echocardiography and CMR L-VEF and no LGE.

References:

[1] Ntusi NA, Piechnik SK, Francis JM, et al. Subclinical myocardial inflammation and diffuse fibrosis are common in systemic sclerosis a clinical study using myocardial T1-mapping and extracellular volume quantification.J Cardiovasc Magn Reson 2014;16:21.
[2] Germain P, El Ghannudi S, Jeung MY et al. Native T1 mapping of the heart - a pictorial review. Clin Med Insights Cardiol 2014;8:1.

Disclosure of Interest: None declared

DOI: 10.1136/annrheumdis-2017-eular.4955

OP0129 MYCOPHENOLATE MOFETIL VERSUS CYCLOPHOSPHAMIDE IN SCLERODERMA-RELATED INTERSTITIAL LUNG DISEASE IN A REAL LIFE SCENARIO

H. Fretheim[1], Ø. Midtvedt[1], E. Volkmann[2], T. Garen[1], M.B. Lund[1], T. Aaløkken[1], Ø. Molberg[1], A.-M. Hoffmann-Vold[1]. *[1]Oslo University Hospital, Oslo, Norway; [2]UCLA, LA, United States*

Background: Systemic sclerosis (SSc) is a serious disease with high risk of interstitial lung disease (ILD); a feared complication with poor prognosis. The SLS 2[1] supported the clinical effectiveness of Mycophenolate Mofetil (MMF) and Cyclophosphamide (Cyc).

Objectives: Evaluate the effect of MMF and Cyc on SSc-ILD in a population based cohort.

Methods: All SSc patients at Oslo University Hospital (OUH) are included in an prospective, observational SSc cohort. All patients are followed annually by rheumatologists and data are recorded in the Norwegian systemic connective tissue disease registry (NOSVAR). Here, we assessed patients diagnosed after the year 2000 (n=433). Longitudinal pulmonary function tests (PFTs), HRCT lung image results, pulmonary hypertension (PH), clinical and demographic data, antibodies and vital status were obtained from NOSVAR. Outcome measures were vital status in December 2016, extent of fibrosis measured on HRCT, FVC and DLCO development.

Results: Among 262 patients with complete data on treatment and ILD, 21 (8%) patients received treatment with Cyc, 14 (5.3%) with MMF and 17 (6.5%) with combination therapy of MMF and Cyc. 55.7% of patients did not receive any treatment for ILD. Baseline characteristics, lung fibrosis and function did not differ significantly between the MMF and Cyc (Table 1). Patients treated with Cyc show a trend of higher mortality. Treatment with Cyc resulted in a significant improvement of lung fibrosis; whereas treatment with MMF resulted in significant progression in lung fibrosis (-1.3% (SD 9.3) and 7.5% (SD 11.1), p=0.024) (Table 1). MMF treatment showed a significant improvement in the DLCO% (1.8% [SD 4.6]); whereas Cyc had a significant decline in the DLCO% (-2.1% [SD 3.5], p=0.14). Less patients treated with MMF developed PH compared with Cyc treatment (1[7.7%] compared to 8[42.1%], p=0.038) (Table 1).

Table 1. Demographics and clinical characteristics of the OUH cohort

	Total cohort (n=262)	Cyc (n=21)	MMF (n=14)	p-value
Age at diagnosis, yrs (SD)	51.4 (15.5)	52.6 (12.1)	43.5 (18.5)	–
Follow-up period, yrs (SD)	6.4 (4.0)	5.5 (3.9)	4.1 (2.3)	–
Male, n (%)	42 (16.0)	6 (28.6)	2 (14.3)	–
dcSSc, n (%)	68 (26.0)	8 (38.1)	10 (71.4)	–
Deceased, n (%)	54 (20.6)	8 (38.1)	4 (28.6)	–
PH, n (%)	35 (14)	8 (42.1)	1 (7.7)	0.038
BL FVC%,(SD)	96.4 (19.5)	83.8 (16.5)	81.5 (13.3)	–
BL DLCO%,(SD)	70.4 (19.5)	54.2 (16.2)	61.8 (14.9)	–
BL fibrosis%,(SD)	6.1 (11.3)	19.1 (15.8)	10.8 (10.3)	–
FVC% decline,(SD)	-0.9 (14.4)	-3.3 (14.3)	2.6 (11.3)	–
Annual FVC% decline,(SD)	-0.02 (6.1)	-0.3 (9.1)	2.3 (4.1)	–
DLCO% decline,(SD)	-9.1 (13.5)	-9 (14.1)	0.7 (12.4)	–
Annual DLCO% decline,(SD)	-2 (5.1)	-2.1 (3.5)	1.8 (4.6)	0.014
Fibrosis% progression,(SD)	1.2 (5.7)	-1.3 (9.3)	7.5 (11.1)	0.024
Annual fibrosis% progression,(SD)	0.5 (2.8)	-0.3 (4.9)	2.3 (4.7)	–

n: number; BL:baseline, SD: standard deviation; FVC: Forced vital capacity; DLCO: diffusing lung capacity for carbon monoxide; dcSSc: diffuse cutaneous SSc; PH: pulmonary hypertension.

Conclusions: Preliminary data from our population based cohort indicate that in a real-life scenario treatment effects of Cyc and MMF appear comparable to randomized clinical trials, but there are some potentially important nuances. Cyc seems to halt fibrosis progression, but toxicity is a major concern, while MMF could have effects on DLCO decline and the development of PH.

References:

[1] Tashkin DP et al. Mycophenolate mofetil versus oral cyclophosphamide in SLS II. Lancet Respir Med. 2016.

Disclosure of Interest: None declared

DOI: 10.1136/annrheumdis-2017-eular.4819